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Assessment of cochlear implant candidacy in patients with cochlear nerve deficiency using the P1 CAEP biomarker

Peter Roland, M.D.,

Department of Otolaryngology Head and Neck Surgery, University of Texas Southwestern Medical Center at Dallas, Dallas, TX

Kathryn Henion, M.A.,

University of Texas Southwestern Medical Center at Dallas, Dallas, TX

Timothy Booth, M.D.,

Department of Pediatric Radiology, University of Texas Southwestern Medical Center at Dallas, Dallas, TX

Julia Dee Campbell, Au.D., and

Department of Speech, Language and Hearing Sciences, University of Colorado at Boulder, Boulder, CO

Anu Sharma, Ph.D

Department of Speech, Language and Hearing Sciences, University of Colorado at Boulder, Boulder, CO

Introduction

Cochlear implantation has become increasingly common in pediatric clinical practice for the treatment of deafness. The prominence of pediatric cochlear implantation since Food and Drug Administration (FDA) approval in 1990 coincides with growing public and professional awareness of the importance of early identification and intervention of hearing loss in children (Francis & Niparko, 2003). While cochlear implantation in children has become a fairly routine procedure, there remain cases where implant candidacy decisions are difficult to make using existing evaluation procedures. Among these cases are children with cochlear nerve deficiencies (CND) (Bradley et al., 2008; Buchman et al., 2006; Govaerts et al., 2003; Walton et al., 2008; Zanetti et al., 2006). Auditory nerve abnormalities or hypoplasias may present in several forms. These forms, including aplasia, have been categorized by Govaerts and colleagues (2003), using magnetic resonance imaging (MRI). Type I consists of a deficient cochleovestibular nerve, with either a normal or dysplastic labyrinth and a stenotic internal auditory canal. Type IIa affects only the cochlear branch of the nerve with labyrinthine dysplasia. The dysplasia may range from mild to a common cavity. Type IIb affects the cochlear branch of the nerve, but the labyrinth is normal.

Compared to other imaging techniques, such as high-resolution computed tomography (HRCT), MRI is considered the gold standard of CND diagnosis (Adunka et al., 2007; Buchman et al., 2006; Lee et al., 2009; McClay et al., 2008; Parry, 2005). However, in some cases, even MRI may not provide enough information to make a decision about information in your child and be limited in determination of an accurate diagnosis of CND.

A viable cochlear nerve is critical to the success of cochlear implantation, because input from the implant needs to be transmitted to the central auditory pathways. Therefore, in cases where the MRI findings are insufficient, it is necessary to determine the integrity of the cochlear nerve using other procedures. Functional assessment such as auditory brainstem

response (ABR) and behavioral audiometry may be useful, and are usually included in any pediatric cochlear implantation battery. However, demonstrating cochlear nerve integrity using these procedures can be confounded by the co-existence of a severe hearing loss and/or auditory neuropathy spectrum disorder (ANSD). For example, severe-to-profound hearing loss (typical of CI candidates) also results in an absent or degraded ABR response, thereby decreasing the utility of the ABR for CNL diagnosis. Furthermore, CNL presents common characteristics with ANSD, such as the presence of a cochlear microphonic in an ABR recording where the ABR is absent, suggesting that similarities between these two pathologies may have to do with the number of functioning neural fibers (Buchman et al., 2006; Walton et al., 2008). In addition, as most pediatric implant recipients are pre-verbal, speech perception testing for evaluating the functional integrity of the central pathways is not possible. Clearly, there exists a need for an objective measure to determine whether stimulation is reaching the central auditory pathways via the cochlear nerve in children with CNL. Such a measure would allow for determination of whether the central pathways in children with CNL would be receptive to cochlear implantation.

Recent studies by Sharma and colleagues (Sharma et al., 2002a), (Sharma et al., 2002b), (Sharma et al., 2002c) (Sharma et al., 2005) have demonstrated the use of cortical auditory evoked potentials (CAEPs) as an objective means of assessing cochlear implant candidacy in infants and young children. In young children, development and plasticity of the central auditory pathways has been studied using the latency and morphology of the P1 CAEP (Ponton et al., 2000; Sharma & Dorman 2006). This component is generated by activity within the auditory cortex including recurrent, cortico-cortical input to the auditory cortex. The P1 is a positive peak occurring at a latency of approximately 50 milliseconds (ms) in adults and 200 ms in newborns. For those with normal hearing, the latency of the P1 CAEP decreases systematically as age increases, reflecting the maturation of the central auditory pathways. Because P1 morphology and latency reliably reflects maturation of auditory cortical pathways, it may be used as a biomarker to assess central auditory development and function in children with hearing loss (Sharma et al., 2005; Sharma & Dorman 2006). The P1 biomarker is easily measured in infants and very young children, as it does not require a behavioral response and is non-invasive. A robust P1 response can be obtained at intensity levels as little as 10 dB SL (above hearing threshold levels), allowing for successful use as a biomarker in children with severe to profound hearing losses (Sharma et al., 2005). Furthermore, as a component of the cortical response, the presence of the P1 requires significantly less synchrony than the ABR (Kraus et al., 1993), which is useful for testing in children with ANSD.

In this report, we assessed the efficacy of the P1 CAEP biomarker as an objective tool to assist in the evaluation of cochlear implant candidacy in four children with severe-to-profound hearing losses who had a radiological diagnosis of CNL. The logic of our approach was as follows: Presence of the P1 biomarker with either normal or delayed latency would suggest that acoustic information relayed from the cochlea was reaching the auditory cortex. Such a patient should be a good candidate for cochlear implantation. On the other hand, an absent P1 response would suggest that the cortical pathways are not being activated by sound, even marginally. In the presence of CNL, such a finding would contraindicate cochlear implantation.

Methods

Subjects

We describe data, in the form of case studies, from four hearing-impaired children: three girls and one boy, tested between the ages of 6 months and 2 years, 3 months. Retrospective review of each patient's chart, imaging studies and CAEP responses was performed. This

research was approved by the Institutional Review Boards of the University of Texas Southwestern Medical Center and The University of Texas at Dallas.

Procedures

Retrospective case study review of electrophysiological (ABR [auditory brainstem response]/OAE [otoacoustic emission]), behavioral audiometric and radiological findings were performed in the four patients identified with cochlear nerve deficiencies.

Stimulus—The P1 CAEP was recorded in response to the synthesized speech syllable /ba/. Details of this stimulus can be found in previously published studies by Sharma et al., 1997; Sharma et al., 2002a; Sharma et al., 2002b; Sharma et al., 2002c. The stimulus was presented via a loudspeaker placed at an angle of 45° to the patient's amplified ear, or using insert earphones if the patient was not aided with amplification at the time of testing. Children who were tested with amplification devices had his/her devices at typical settings during these measures. Subjects were seated comfortably in a reclining chair placed in a sound booth. Subjects watched a videotape movie or cartoon of their choice on a TV monitor placed in front of them in the sound booth. Videotape audio levels were kept to a minimum volume level (below 45 dB SPL).

Data Analysis—Evoked potentials were collected using a Compumedics Neuroscan® evoked potential system. Silver/silver chloride cup electrodes were used for the recordings, with Cz as the active electrode. Cz refers to the vertex midline placement, as in the International 10–20 System. The reference electrodes were placed on the mastoids and a ground electrode on the forehead. Eye movements were monitored using a bipolar electrode montage (lateral outer canthus-superior outer canthus). Averaging was automatically suspended by the recording computer when eye blinks were detected. The recording window included a 100 ms pre-stimulus time and 600 ms post-stimulus time. Incoming evoked responses were analog filtered from 0.1 Hz to 100 Hz. Approximately two runs of 300 response sweeps were collected for each subject. The test session, including electrode application and evoked response recording, lasted for about 30 minutes. Sweeps greater than $\pm 100\mu\text{V}$ were rejected off-line and the remaining sweeps were averaged to compute a grand average waveform for the individual subjects. The P1 was defined as the first robust positivity in the waveform. The P1 latency was labeled at the peak of the response or, if the peak was broad, at the midpoint. P1 response latencies were plotted against the 95% confidence interval for normal development of the P1 response (Sharma et al., 2002c).

Results

Case 1

Patient 1 was a male child who was diagnosed via ABR at 21 months of age as having ANSD (Starr et al., 1996; Berlin et al., 1998). No significant medical history was reported for this patient. Magnetic resonance imaging (MRI) studies showed absence of both cochlear nerves with mild dysplasia of both cochleas. The internal auditory canals were reported as being normal in size; however, marked hypoplasia of the cochlear nerve canal was observed (Fig. 1). Behavioral audiometric testing had not been completed for this patient at the time of this referral for P1 CAEP measures.

Individual ear P1 responses using insert earphones were recorded at age 25 months (Fig. 2a). P1 responses were clearly present and robust. However, the P1 latencies were delayed in both ears compared to the 95% confidence interval for normal development of the P1 response (Sharma et al., 2002c). Delayed P1 responses suggested that he was not receiving adequate auditory input for the development of the central auditory pathways. Based on the

presence of well-formed P1 responses, the patient was referred for comprehensive behavioral audiometric testing and a trial with amplification. Unaided behavioral thresholds were obtained in the severe-to-profound hearing loss range bilaterally (pure-tone averages [PTA] = 88 dB HL, right ear; 91 dB HL, left ear) (Fig. 2b). As a result of behavioral testing, the patient was fit with binaural hearing aids at age 29 months. Behavioral aided testing via soundfield using binaural hearing aids revealed a pure-tone average (at 0.5 and 1 kHz) of 65 dB HL (Fig. 2b). Based on ABR, P1, and behavioral audiometric results, a recommendation was made to consider cochlear implants for this patient. At present, the child is completing all pre-implantation evaluations.

In this case of ANSD, in which the ABR and MRI were abnormal, the presence of robust bilateral P1 responses indicated that acoustic information was activating the central auditory pathways, albeit insufficiently. The delayed P1 latencies were consistent with the severe-to-profound degree of hearing impairment, confirmed by behavioral audiometry. Thus, the P1 results, in conjunction with behavioral audiometry, provided useful information resulting in a recommendation for cochlear implantation.

Case 2

Patient 2, a female child, had a complicated peri-natal history, including hypoxia, left brachial plexus injury and mechanical ventilation. ABR testing at age 22 months estimated a severe-to-profound sensorineural hearing loss, bilaterally. Retrospective review of ABR tracings for this patient revealed electrophysiologic characteristics of ANSD (i.e., presence of cochlear microphonic in ABR recordings) for the right ear only (Starr et al., 1996; Berlin et al., 1998), though a formal diagnosis of ANSD was never made in this case. MRI findings revealed mild cochlear dysplasia with loss of the modiolus bilaterally; a definitive cochlear nerve could not be visualized on either side. The right vestibulocochlear complex was absent and the left was very small (Fig. 3). Results of audiometric testing after the patient had been fitted with hearing aids at age 27 months are shown in Figure 4b. Results revealed unaided thresholds in the severe-to-profound hearing loss range for the left ear (PTA= 96 dB HL), and no observable response at any test frequency for the right ear (Fig. 4b). Aided testing revealed a PTA of 65 dB HL for the left ear, while no observable aided response for the right ear was noted at any test frequency.

The ear-specific aided P1 responses were recorded at 1 and 6 months post-hearing aid fitting. Results for the 6-month point are shown in Figure 4a. A robust P1 CAEP response was observed for the patient's left ear, with no replicable P1 response evident for the right ear (Fig. 4a). However, the P1 latency in the left ear was delayed when compared to age-appropriate 95% confidence intervals (Sharma et al., 2005). Based upon the audiometric data and presence of a delayed P1, the recommendation was made to implant the left ear and surgery was performed at age 3 years. In this case, the unilateral presence of the delayed P1 response assisted in the decision of ear selection for implantation.

Case 3

Patient 3, a female child, underwent ABR testing at age 2 months, which estimated the presence of a severe-to-profound hearing loss for the right ear and a severe hearing loss for the left ear. Behavioral audiometric testing was attempted post-identification at age 5 months, albeit with limited success given the patient's young test age. Based on the estimated ABR result, the patient was fit with binaural hearing aids at age 5 months. MRI was completed for this patient at age 11 months. MRI findings revealed absent cochlear nerves and cochlear nerve canals bilaterally (Fig 5). P1 CAEP testing was performed at age 6 months, one month post-hearing aid fitting. Aided P1 results revealed a robust and replicable response (Fig. 6a) with normally developing morphology and latency bilaterally

when compared to age-appropriate 95% confidence intervals (Sharma et al., 2005). Due to P1 results that suggested a normally developing central auditory system, it was requested that the ABR and behavioral testing be repeated. A second ABR completed at age 8 months yielded the same result as initial testing. Reliable behavioral testing using visual reinforcement audiometry was completed for this patient at 9 months of age. Testing revealed unaided pure-tone thresholds in the moderate hearing loss range (PTAs =52 dB HL left ear; 55 dB HL right ear), with an aided PTA of 33 dB HL (Fig. 6b). Behavioral testing indicated hearing thresholds better than originally predicted by ABR, which is not uncommon (though usually not to this degree). Aided behavioral hearing thresholds were consistent with the P1 latencies, suggesting that amplification was providing adequate input for normal development of the central auditory pathways. P1 latencies continued to remain normal for this patient.

In this case of a child with reported absence of cochlear nerves and canals, P1 testing at a very early age suggested that the central auditory pathways were adequately stimulated via the auditory nerve and maturing normally. Aided behavioral testing (performed when the child was older) confirmed these results. At present the child is performing well with her hearing aids and we continue to monitor her development.

Case 4

Patient 4, a female child, failed her newborn hearing screening and was referred for further audiological assessment. No significant birth or medical history was reported. Behavioral audiometric testing, conducted in soundfield for this patient at age 11 months, suggested a profound hearing loss with no observable response to any test frequency. A profound bilateral hearing loss was also estimated using ABR measures at age 12 months. Her MRI revealed severe cochlear and vestibular dysplasia with absent cochlear nerves bilaterally (Fig. 7).

This child was fitted bilaterally with hearing aids; however, aided audiometric testing revealed no observable responses to any test frequency for either ear (Fig. 8a). P1 CAEP testing also revealed absent, non-replicable P1 responses when tested in a binaurally aided condition at high intensity levels (~95 dB HL) (Fig. 8b). This finding is consistent with the audiological profile for this child, as children with profound hearing loss (in this case, no response even with acoustic amplification) are unlikely to show activation of the central auditory pathways and primary auditory cortex due to lack of stimulation (Sharma et al., 2005). Though not ear-specific, even a P1 resulting from a binaural condition should be sufficient for normal speech and language acquisition (Nash et al., 2008). Thus, the lack of a P1 response indicates insufficient auditory access to speech and language. Behavioral audiometric testing was repeated at several follow-up visits for this patient and results continued to demonstrate no response to any test frequency for either ear.

Based on the collective evidence of MRI, behavioral, and P1 CAEP test results, cochlear implantation was not recommended for this patient. A recommendation was made to pursue a manual mode of communication as a primary means for language input.

In this case, behavioral audiometric testing demonstrated that the child was non-responsive to sound, even with hearing aids. Absent P1 responses at very high intensity levels indicated that no acoustic information was being relayed to the central auditory pathways, resulting in an absence of stimulation of these pathways and primary auditory cortex. These results, taken together, are consistent with the radiological diagnosis of cochlear dysplasia and the absence of cochlear nerves bilaterally.

Discussion

In this study, we explored the utility of the P1 CAEP biomarker in assessing cochlear implant candidacy in children with CND. At issue was whether the P1 response would be a reliable indicator of central auditory function and development in children with severe-to-profound hearing loss who are diagnosed with CND.

Three out of four children reported in this study demonstrated robust P1 responses, indicating that their central auditory pathways were being activated by auditory stimulation. This activation had to occur via the cochlear nerve, despite the diagnosis of CND. In two children (Cases 1 & 2), the P1 response was delayed relative to their normal-hearing age-matched peers, consistent with the diagnosis of severe-to-profound degree of hearing loss. Case 3, with a moderate hearing loss, showed normal central auditory development (as reflected by age-appropriate P1 latencies) after 6 months of hearing aid use. Interestingly, in this sample of four children with CND, one child had bilateral ANSD (Case 1) and another child likely had unilateral ANSD (Case 2). This finding is consistent with the relatively high rate of ANSD seen in children with CND (Buchman et al., 2006; Walton et al., 2008). The presence of robust cortical responses is typically associated with good behavioral outcomes, though greater research is required in the area of cortical measures and ANSD (Cone-Wesson & Wunderlich, 2003; Rance et al., 2002). Cochlear implantation of children with ANSD is not conclusive in regards to success, with mixed reports present in the literature (Peterson et al., 2003; Walton et al., 2008; Zanetti et al., 2006). Such inconclusiveness concerning cochlear implant success in children with ANSD may simply be due to the lack of a routine CND evaluation for the population, as CND may present with ANSD characteristics (Buchman et al., 2006; Laury et al., 2009; Walton et al., 2008). Overall, in Cases 1–3, all of whom showed abnormal ABR responses, the presence of the P1 was a reliable indicator of central auditory functioning and assisted in the decision of whether (and for which ear) to provide the child with a cochlear implant.

Finally, we were not able to obtain P1 responses in only one child (of four) described in this paper. Case 4 showed absent P1 responses, even when tested at high intensity levels. The absent P1 responses were consistent with the child's profound hearing loss as documented using behavioral audiometry. In this case, the profound hearing loss, documented by the P1 and behavioral audiometry testing, were taken to be consistent with MRI results of cochlear hypoplasia and cochlear nerve hypoplasia. Based on the electrophysiological, behavioral and radiological findings, we determined that this child would not benefit from a cochlear implant. In making this decision, the assumption was made that the profound hearing loss resulted, at least in part, from CND. For this case of CND, additional testing may have been useful in order to ascertain whether central pathways would respond to electrical stimulation from an implant. Walton et al. (2008) and Kileny and Zwolan (2004) both describe pre-operative trans-tympanic or promontory electrically evoked auditory brainstem response (EABR) measures as an effective tool to provide quantitative information regarding residual auditory innervation. It may be of interest to combine P1 testing with cochlear electrical stimulation in patients with CND to determine prognostic value and utility in evaluating the presence of cochlear neural fibers.

As we have described in this study, we felt that Cases 1 and 2 were good candidates for implantation, however, we do not have post-implantation outcomes in these children at this time. Bradley et al. (2008) reports six case studies of children diagnosed with VIII nerve hypoplasia and who received a CI. The child with the largest vestibulocochlear nerve performed the best of group; the authors report that at 3 years post-implantation he was able to understand common phrases without lipreading. The other cases, including those who were implanted early, relied more on manual communication. Govaerts et al. (2003) reports

similar findings with four case studies. All four children preferred total communication settings though one attended an oral educational setting. Zanetti and colleagues (2006) report a single case study in which the subject underwent early implantation for hypoplasia. While the authors report that the child benefits greatly from both a CI and hearing aid, it is also stated that he would not be able to function auditorily with his CI alone. Finally, Walton et al. (2009) evaluated CI outcomes in children with ANSD, split into two groups: one comprised of children with CND, and one with normal cochlear nerves. Both groups had a similar mean implantation age of approximately 44 and 40 months. Overall, the children with normal cochlear nerves and ANSD outperformed children with ANSD and CND on the Melbourne Speech Perception Score (MSPS). Therefore, while children with CND may clearly benefit from an implant, the amount of benefit may depend on the magnitude and viability of the cochlear nerve. These studies pose the question, 'how much of an auditory nerve is necessary for optimal speech perception and for a normal P1 to be obtained?' We aim to answer this question in future studies. For these presented cases, utilization of the P1 CAEP measure in conjunction with MRI measurements of the VIII nerve and speech perception testing provided information to evaluate CI candidacy. Once the children described here are able to perform speech perception tasks, we will document their performance to add to the literature on pediatric CND and cochlear implants.

This study emphasizes the importance of an efficient and sensitive test battery, including MRI, ABR, and behavioral audiometry, in order to successfully determine CI candidacy for children with CND. The P1 evaluation of central auditory function and maturation would be a valuable addition to such a battery.

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References

- Adunka OF, Jewells V, Buchman CA. Value of computed tomography in the evaluation of children with cochlear nerve deficiency. *Otol Neurotol*. 2007; 28(5):597–604. [PubMed: 17667769]
- Berlin CI, Bordelon J, St John P, Wilensky D, Hurley A, Kluka E, Hood LJ. Reversing click polarity may uncover auditory neuropathy in infants. *Ear Hear*. 1998; 19(1):37–47. [PubMed: 9504271]
- Bradley J, Beale T, Graham J, Bell M. Variable long-term outcomes from Cochlear implantation in children with hypoplastic auditory nerves. *Cochlear Implants Int*. 2008; 9(1):34–60. [PubMed: 18246534]
- Buchman CA, Roush PA, Teagle HF, Brown CJ, Zdanski CJ, Grose JH. Auditory neuropathy characteristics in children with cochlear nerve deficiency. *Ear Hear*. 2006; 27(4):399–408. [PubMed: 16825889]
- Cone-Wesson B, Wunderlich J. Auditory evoked potentials from the cortex: Audiology applications. *Curr Opin Otolaryngol Head Neck Surg*. 2003; 11(5):372–377. [PubMed: 14502069]
- Francis HW, Niparko JK. Cochlear implantation update. *Pediatr Clin North Am*. 2003; 50(2):341–361. viii. [PubMed: 12809327]
- Govaerts PJ, Casselman J, Daemers K, De Beukelaer C, Yperman M, De Ceulaer G. Cochlear implants in aplasia and hypoplasia of the cochleovestibular nerve. *Otol Neurotol*. 2003; 24(6):887–891. [PubMed: 14600469]
- Kileny PR, Zwolan TA. Perioperative, transtympanic electric abr in paediatric cochlear implant candidates. *Cochlear Implants Int*. 2004; 5(Suppl 1):23–25. [PubMed: 18792220]
- Kraus N, McGee T, Ferre J, Hoeppe J-A, Carrell T, Sharma A, Nicol T. Mismatch negativity in the neurophysiologic/behavioral evaluation of auditory processing deficits: A case study. *Ear Hear*. 1993; 14(4):223–234. [PubMed: 8405727]
- Laury AM, Casey S, McKay S, Germiller JA. Etiology of unilateral neural hearing loss in children. *Int J Pediatr Otorhinolaryngol*. 2009; 73(3):417–427. [PubMed: 19155072]

- Lee SY, Cha S-H, Jeon MH, Bae IH, Han GS, Kim SJ, Park KS. Narrow duplicated or triplicated internal auditory canal (3 cases and review of literature): Can we regard the separated narrow internal auditory canal as the presence of vestibulocochlear nerve fibers? *J Comput Assist Tomogr.* 2009; 33:565–570. [PubMed: 19638851]
- McClay JE, Booth TN, Parry DA, Johnson R, Roland P. Evaluation of pediatric sensorineural hearing loss with magnetic resonance imaging. *Arch Otolaryngol Head Neck Surg.* 2008; 134(9):945–952. [PubMed: 18794439]
- Nash, A.; Sharma, A.; Martin, K.; Biever, A. Clinical applications of the P1 cortical auditory evoked potential (CAEP) biomarker. *A Sound Foundation Through Early Amplification: Proceedings of a Fourth International Conference; 2007; Chicago, IL.* 2008.
- Nikolopoulos TP, Mason SM, Gibbin KP, O'Donoghue GM. The prognostic value of promontory electric auditory brainstem response in pediatric cochlear implantation. *Ear Hear.* 2000; 21(3): 236–241. [PubMed: 10890732]
- Parry DA, Booth T, Roland PS. Advantages of magnetic resonance imaging over computed tomography in preoperative evaluation of pediatric cochlear implant candidates. *Otol Neurotol.* 2005; 26(4):976–982. [PubMed: 16151346]
- Peterson A, Shallop J, Driscoll C, Breneman A, Babb J, Stoeckel R, Fabry L. Outcomes of cochlear implantation in children with auditory neuropathy. *J Am Acad Audiol.* 2003; 14(4):188–201. [PubMed: 12940703]
- Ponton CW, Eggermont JJ, Kwong B, Don M. Maturation of human central auditory system activity: evidence from multi-channel evoked potentials. *Clin Neurophysiol.* 2000; 111:220–236. [PubMed: 10680557]
- Rance G, Cone-Wesson B, Wunderlich J, Dowell R. Speech perception and cortical event related potentials in children with auditory neuropathy. *Ear Hear.* 2002; 23(3):239–253. [PubMed: 12072616]
- Sharma A, Dorman MF. Central auditory development in children with cochlear implants: Clinical implications. *Adv Otorhinolaryngol.* 2006; 64:66–88. [PubMed: 16891837]
- Sharma A, Dorman MF, Spahr AJ. Rapid development of cortical auditory evoked potentials after early cochlear implantation. *Neuroreport.* 2002b; 13(10):1365–1368. [PubMed: 12151804]
- Sharma A, Dorman MF, Spahr AJ. A sensitive period for the development of the central auditory system in children with cochlear implants: implications for age of implantation. *Ear Hear.* 2002c; 23(6):532–539. [PubMed: 12476090]
- Sharma A, Dorman M, Spahr A, Todd NW. Early cochlear implantation in children allows normal development of the central auditory pathways. *Ann Otol Rhinol Laryngol Suppl.* 2002a; 189:38–41. [PubMed: 12018346]
- Sharma A, Kraus N, McGee TJ, Nicol TG. Developmental changes in P1 and N1 central auditory responses elicited by consonant-vowel syllables. *Electroencephalogr Clin Neurophysiol.* 1997; 104(6):540–545. [PubMed: 9402896]
- Sharma A, Martin K, Roland P, Bauer P, Sweeney MH, Gilley P, Dorman M. P1 latency as a biomarker for central auditory development in children with hearing impairment. *J Am Acad Audiol.* 2005; 16(8):564–573. [PubMed: 16295243]
- Starr A, Picton TW, Sininger Y, Hood LJ, Berlin CI. Auditory neuropathy. *Brain.* 1996; 119((Pt 3)): 741–753. [PubMed: 8673487]
- Walton J, Gibson WP, Sanli H, Prelog K. Predicting cochlear implant outcomes in children with auditory neuropathy. *Otol Neurotol.* 2008; 29(3):302–309. [PubMed: 18317399]
- Zanetti D, Guida M, Barezzani MG, Campovecchi C, Nassif N, Pinelli L, Giordano L, Oliosio G. Favorable outcome of cochlear implant in VIIIth nerve deficiency. *Otol Neurotol.* 2006; 27(6): 815–823. [PubMed: 16936567]

a.

b.

c.

Figure 1.

1a, 1b, 1c.

For Case 1: 1a shows a 3D axial T2 weighted image through the cochlea. The modiolus is flattened on both sides consistent with mid dysplasia. The cochlear nerve canal is narrowed on the left and not visualized on the right (arrow).

1b (left) and 1c (right) show a 3D oblique sagittal T2 weighted images through the mid IAC. No cochlear nerve is identified on either side. A normal facial nerve and vestibular complex is present (long arrows).

Figure 2.

2a, 2b.

2a shows P1 response waveforms for Case 1 obtained for left ear stimulation (top waveform) and right ear stimulation via insert earphones (bottom waveform). P1 responses were present and well-formed, albeit delayed. For all CAEP figures, amplitude is in microvolts, with positive polarity displayed upward. Latency is shown on the abscissa in milliseconds.

2b shows the behavioral audiometric information for hearing threshold levels. For all audiometric figures, X represents left ear responses, O represents right ear responses. A represents binaural aided responses. Intensity level in dB HL is displayed on the ordinate

and frequency in hertz on the abscissa. For Case 1, aided responses show a corner response in the severe range, unaided responses in the profound range.

a.

b.

c.

d.

Figure 3.

3a, 3b, 3c, 3d.

For Case 2: 3a and 3b show a 3D axial T2 weighted images through the cochlea. There is decreased partition of the cochlea with a deficient modiolus. The internal auditory canals are markedly hypoplastic.

3c (left) and 3d (right) show a 3D oblique sagittal images through the medial IAC/cerebellopontine cistern. The facial nerve was seen bilaterally (arrow). The normal vestibulocochlear complex is 2.5 times the diameter of the facial nerve. No vestibulocochlear complex is seen on the right and a small complex is present on the left (long arrow).

Figure 4.

4a, 4b.

4a shows P1 response waveforms for Case 2 for the left aided (HA) ear (top waveform) and the right aided ear (bottom waveform). The P1 response in the left aided ear is robust and well-formed, though delayed. The right aided ear shows no replicable P1 response.

4b shows the behavioral audiometric information for hearing threshold levels for Case 2. L represents left aided responses, R right aided responses. The left shows an aided response in the severe range and an unaided response in the severe-profound range. The right aided and unaided responses are in the profound range with no response at output limits of the audiometer (note arrows on symbols).

a.

b.

C.

Figure 5.

5a, 5b, 5c, 5d.

For Case 3: 3a and 3b show a 3D axial T2 weighted images through the cochlea. Bone is present within the expected location of the cochlear nerve canal (arrow).

3c (left) and 3d (right) show a 3D oblique sagittal T2 weighted images through the mid IAC. No definite cochlear nerve is demonstrated on either side. The facial nerve and vestibular complex are present (long arrow).

Figure 6.

6a, 6b.

6a shows P1 response waveforms for Case 3 for left ear stimulation (top waveform) and right ear stimulation via insert earphones (bottom waveform). Both ears demonstrate robust and well-formed P1 responses with age-appropriate latencies.

6b shows the behavioral audiometric information for Case 3. Aided responses show a response in the moderate range, unaided responses in the moderate-severe range.

a.

b.

Figure 7.

7a, 7b, 7c.

For Case 4: 7a shows a 3D axial T2 weighted image through the membranous labyrinth. Severe cochlear and vestibular dysplasia is present bilaterally. A single cochlear turn is present on both sides (arrow).

7b and 7c shows a 3D oblique sagittal T2 weighted images through the mid IAC. The cochlear nerves are not visualized. The facial nerve and vestibular complex is present (long arrows).

Figure 8.

8a, 8b.

8a shows the P1 response waveform for a binaural aided condition for Case 4. In this case, the P1 response is absent, with no peak present.

8b shows the behavioral audiometric information for hearing threshold levels for Case 4. No response is present for any condition at the output limits of the audiometer (note arrows on symbols).